A mentally retarded man with a nasal discharge

Peter K M Ku, John N Marshall, Charles Andrew van Hasselt

A 35-year-old mentally retarded Chinese man presented with a 6 month history of intermittent, foul-smelling nasal discharge from his left nostril. He had been treated with antibiotics, topical nasal decongestants and steroid nasal sprays without success, and plain sinus X-rays had shown an opaque left maxillary antrum. Because of this a left antral wash-out had been performed on two occasions, followed by an inferior meatal antrostomy, but still without resolution of his symptoms. His medical history included glaucoma affecting the left eye for which he had undergone surgery, and a history of grand-mal epilepsy treated with anticonvulsants. Examination revealed port-wine naevi over his left frontal and malar regions, and hypertrophy of the frontal and maxillary bone on the left. Purulent secretions were present in the left nasal cavity. The nasal mucosa was congested but no polyps, foreign bodies or other obstructive lesions were seen. Neurological examination revealed a reduction of the motor power of his right arm and leg to grade 4/6. In view of his persistent nasal symptoms, a computed tomography (CT) scan of the paranasal sinuses was performed (figures 1–3).

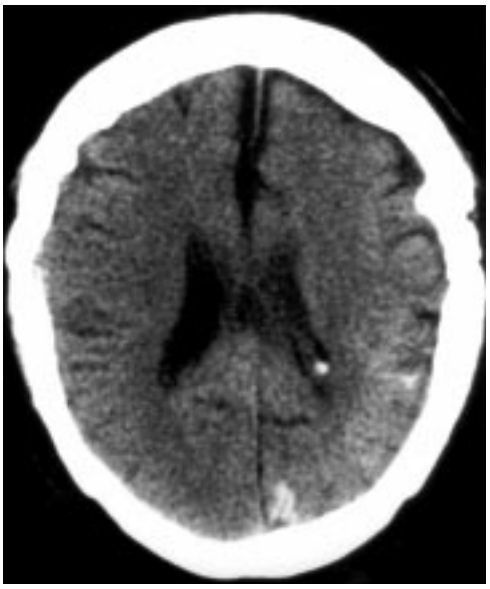

Figure 1 Axial contrast CT brain scan

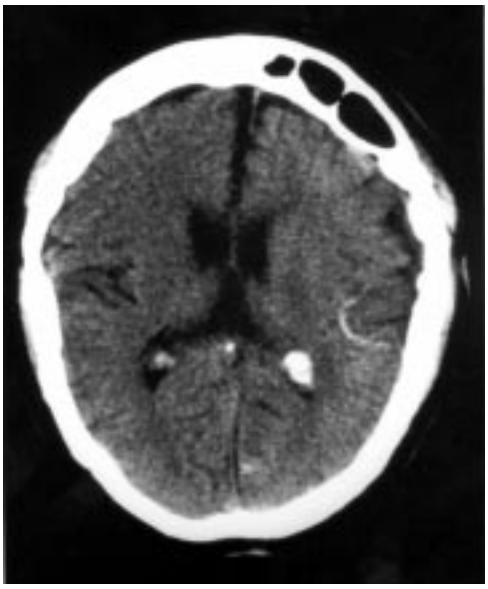

Figure 2 Axial contrast CT brain scan

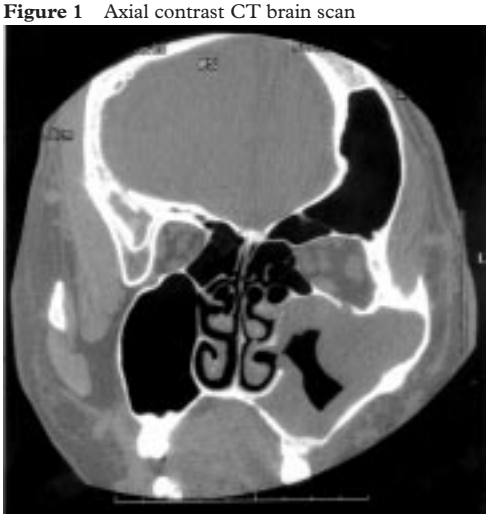

Figure 3 Coronal non-contrast CT brain scan

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Questions

1 List the radiological abnormalities visible on the CT scans.
2 What is the most likely congenital syndrome that this man is suffering from and what is the differential diagnosis?
3 What is the most likely diagnosis of his nasal problem and what is the treatment?

Answers

QUESTION 1

Figure 1 shows cerebral atrophy and, tram-line cortical calcification of the left occipital region Figure 2 shows hypertrophy of the left choroid plexus in the left ventricle, and tram-line calcification in the left occipital and the parietal region of the brain cortex. On the coronal non-contrast CT scan (figure 3), mucosal thickening of the left maxillary sinus, hypertrophy of the frontal and maxillary sinuses, a prominent frontal diploeic bone, and evidence of previous left inferior meatal antrostomy can be seen.

QUESTION 2

The diagnosis is encephalotrigeminal angiomatosis (Sturge-Weber syndrome). The differential diagnosis includes Wyburn-Manson syndrome and leukodystrophy with meningeal angiomatosis.

QUESTION 3

This patient has been suffering from chronic maxillary sinusitis. As conservative treatment by antibiotics and steroids had failed and the symptoms recurred after antral wash-out, functional endoscopic sinus surgery is indicated. A middle meatal antrostomy is required under endoscopic guidance. This improves ventilation and restores normal mucociliary clearance towards the natural ostium of the maxillary sinus.

Discussion

This patient was diagnosed as having encephalotrigeminal angiomatosis (Sturge-Weber syndrome) during childhood. It is a non-hereditary condition that includes a port-wine capillary naevus on the face and is often in the distribution of the first division of the trigeminal nerve. These patients may also have convulsions which are focal and involve the contralateral side of the body. Contralateral hemiparesis and mental retardation may be present. Occasionally homonymous hemianopia is reported. Ipsilateral intracranial calcifications are found on plain skull X-ray. These calcifications are characteristically in paired lines, termed 'tram-line' calcification. Increased intra-ocular pressure may be caused by angiomatous involvement of the uveal tract and thus can give rise to enlargement of the involved globe. About two-thirds of patients with this condition have epilepsy. There is great variability in the severity of the individual symptoms, and one or more may be missing entirely. Haemangiomas may be found in parts of the body other than the face, but rarely in the fundi. These may be extensive and associated with hypertrophy of the limbs and deep varices.

The intracranial lesion is a capillary haemangioma that involves the meninges in the area supplied by the first division of the trigeminal nerve. It is related to the superficial vessels occupying the sulci over the convexity, particularly in the occipital and parietal regions. Such haemangiomas may cause atrophy of the underlying brain tissue. Degenerative changes in cerebral tissue just below the gyral surface cause the characteristic calcifications which are limited to the convexity of the brain. The characteristic gyral pattern is almost pathognomonic of this condition.

Radiological diagnosis of Sturge-Weber syndrome is based on the recognition of the characteristic 'tram-line' calcifications on plain skull X-ray. However, CT scanning makes it possible to identify the classical signs of Sturge-Weber syndrome earlier and more clearly than on a plain X-ray, by allowing early recognition of small intracranial calcifications, cerebral atrophy, unilateral shrinkage of the cranium, hypertrophy of diploeic bone or paranasal sinus hypertrophy. Moreover, anomalies of vasculature such as pial angiomatosis or choroid plexus hypertrophy which have only been identifiable on angiography in the past[1] can be demonstrated clearly on a CT scan. More recently, magnetic resonance imaging has been found to be more efficient in making the diagnosis and detecting lesions related to the clinical neurological status of the patient.[2]

Therapy is symptomatic, although early surgical excision in the form of lobectomy and hemispherectomy is sometime done in the hope of preventing seizures that may be difficult to control and associated with intellectual decline.[3] The treatment option for chronic maxillary sinusitis is either medical or surgical.[4] Once a dental cause for the unilateral maxillary sinusitis has been excluded, a minimum course of 10 days of broad-spectrum oral antibiotics which covers both aerobic and anaerobic infection should be given. The addition of a short course of topical nasal steroids may help to reduce mucosal swelling around the sinus orifice and regain mucociliary clearance of the sinus. If conservative treatment fails, a sinus washout can be performed after 3 to 4 weeks to clear the mucopus collected in the sinus. Pus is thus obtained from the sinus for culture and sensitivity which help to guide the antibiotic treatment. More than one antral washout may be required before symptoms can effectively be alleviated. However, if the above means fail to control symptoms, a sinus-drainage operation is required. In the past, inferior meatal antrostomy and the 'Caldwell-Luc' operation were the mainstays of surgical treatment for chronic maxillary sinusitis. These aimed to restore the ventilation, allow free drainage of secretions by gravity and eradicate diseased mucosa in the antrum. Recent research on chronic sinusitis has stressed the importance of the ethmoid–middle meatal complex, an area where the mucociliary clearance of secretions from the frontal, ethmoid and maxillary sinuses may be obstructed. Functional endoscopic sinus surgery (FESS), based on logical physiological concepts, has addressed these underlying problems by aiming to restore ventilation and mucociliary clearance of the paranasal sinuses.[5] Using rigid endoscopes of different viewing angles (0^0, 30^0 and 70^0), allows excellent visualisation of the hidden areas within the nasal cavities and paranasal sinuses. FESS is less traumatic than traditional surgery and requires shorter duration of hospitalisation.

Final diagnosis

Encephalotrigeminal angiomatosis (Sturge-Weber syndrome).

1 Fotter R, Sager WD, Schneider GH, *et al*. Computer tomography in the Sturge-Weber syndrome. *Computertomographie* 1981;**1**:80–2.
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3 Holmes GL. Surgery for intractable seizures in infancy and early children. *Neurology* 1993;**43**:S28–S37.
4 Mackay I.Infective rhinitis and sinusitis. In: *Scott-Brown's Otolaryngology,* 6th edn, vol 3. Butterworths, London, 1997; pp 810–35.
5 Stammberger H. Endoscopic endonasal sinus surgery. Concept in treating of recurrent rhinosinusitis. Part I. Anatomic and pathophysiologic considerations. *Otolaryngol Head Neck Surg* 1986;**94**:143–7.

Keywords: encephalotrigeminal angiomatosis; Sturge-Weber syndrome; nasal discharge

Sudden-onset watery diarrhoea in a middle-aged woman

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A 55-year-old woman presented with a 2-month history of profuse watery diarrhoea. The onset of diarrhoea was very sudden – she was woken up at 04.00 h one morning with a severe urge to move her bowels. The motions were watery, frequency averaging five times per day. There was no rectal bleeding, abdominal pain or vomiting. She had lost 2 kg in weight but there was no history of iritis, arthritis, rashes or aphthous ulcers. Clinical examination was unremarkable. Laboratory investigations revealed normal full blood count, urea and electrolytes. C-Reactive protein was not raised and there was no growth on stool culture. Colonoscopy showed mild erythema with loss of vascular pattern in the transverse colon, but no ulceration. Multiple biopsies were taken from the region. Histology of the colonic biopsy is shown in the figure.

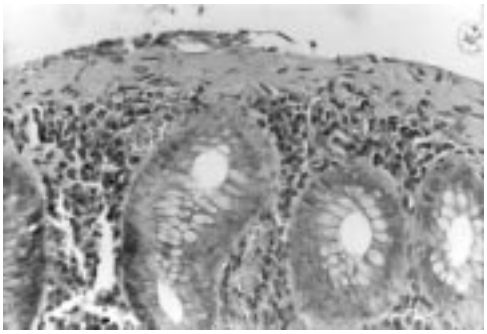

Figure Histology of the colonic biopsy

Questions

1 What is the diagnosis?
2 What is the pathogenesis of this condition?

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